Spontaneous Intracranial Hypotension in a Patient with Reversible Pachymeningeal Enhancement and Brain Descent

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A 56-year-old woman presented with severe orthostatic headache in association with nausea and vomiting. Lumbar puncture for the patient revealed significantly low cerebrospinal fluid pressure (CSF) and the clinical diagnosis of intracranial hypotension syndrome was made. An initial gadolinium-enhanced brain magnetic resonance imaging (MRI) disclosed diffuse meningeal enhancement as well as brain sagging. No definite CSF leakage was found using radionuclide cisternography. Her headaches abated with proper usage of analgesics, strict bed rest, and intravenous hydration. Follow-up neuroimaging studies showed partially resolved meningeal enhancement 2 months after treatment and complete resolution 6 months after treatment. The temporal changes found on MRI suggest that the pachymeningeal enhancement is reversible in patients with spontaneous intracranial hypotension. Moreover, proliferation of meninges is likely to be responsible for this type of delayed resolution phenomenon. *(Chang Gung Med J 2003;26:293-8)*

Key words: intracranial hypotension, orthostatic headache, cerebrospinal fluid, magnetic resonance imaging.

Intracranial hypotension (IH) is a syndrome characterized by postural related headaches and low cerebrospinal fluid (CSF) pressure. Clinically, it is often accompanied by nausea, vomiting, and sometimes photophobia, tinnitus, vertigo, blurred vision, or diplopia.[1-4] The IH syndrome can be divided into two categories including the spontaneous type and symptomatic type. The former is diagnosed with no evidence of CSF leakage while the latter is associated with definite CSF leakage.[5] Moreover, the diagnosis of spontaneous IH should be restricted to patients with no prodromal central nervous system trauma or prior lumbar puncture. In addition, the existence of a dural tear should be carefully ruled out before the diagnosis is made.[3-5] Apart from the radionuclide cisternography, magnetic resonance imaging (MRI) with gadolinium enhancement has been very helpful in the differential diagnosis of this entity.[4,6,7] We herein describe a patient with typical clinical manifestation in association with temporal changes of pachymeningeal enhancement and brain descent evidenced by a series of follow-up MRI studies.

CASE REPORT

A 56-year-old housewife suffered from severe headaches in December of 1999. At first, she experienced a dull head pain when she got up from her bed. The pain was mainly located at the vertex portion in association with tightness of the occipital and postnuchal areas. In particular, the headache was

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precipitated by erect posture and ameliorated immediately on lying flat. Dizziness, nausea, vomiting and tinnitus were also symptoms once the headaches became severe. She took acetaminophen and other non-steroid-antiinflammatory-drugs (NSAIDs) suggested by pharmacists and local clinicians for relief of the headaches. However, her headaches became incrementally more intense during the 3 weeks prior to admission and she was incapable of doing her housework. She denied any history of cranial or spinal trauma.

The patient was admitted to our ward in January of 2000 for investigation of the incapacitating headaches. The results of her complete physical examination were normal. On neurological examination, she was alert and oriented. Examination results of cranial nerve and eye fundus were unremarkable. No evidence of motor or sensory deficits were found. An elicitation of meningeal irritation sign was negative. An initial computed tomographic (CT) scan of the brain revealed slit-like ventricles and prominent meningeal enhancement. A CSF study revealed low opening CSF pressure (30 mm CSF), mild pleocytosis (WBC: 8 cells$\times 10^9$/L with mononuclear cells dominant), normal glucose level, and increased protein content (1150 mg/L). No subarachnoid block was found via the Queckenstedt test method. Cultures and cytological examination results of CSF were all negative. Accordingly, a clinical diagnosis of intracranial hypotension was established. Brain MRI with gadolinium enhancement showed bilateral subdural effusion as well as diffuse meningeal enhancement (Fig. 1A). In addition, downward displacements of the corpus callosum and cerebellar tonsil were disclosed on sagittal T1-weighted imaging (Fig. 1B). For further search of possible CSF leakage, a retrograde radionuclide cisternography was performed via a lumbar puncture using 10 μCi Technetium-99m-diethylamine-triamine-penta-acetic acid. The patient was in supine posture with scintigrams taken at 1, 2, 5, 24 and 48 hours, respectively. In addition to the slow ascent of the radioisotope along the spinal axis, non-visualization of radioactivity in the lateral ventricles was also found in serial scans. In the cerebral convexities, relative delayed and less than expected radioactivities were seen on the 24-hour scan. Of special note, no evidence of CSF leakage was identified.

Her headaches aggravated after the lumbar puncture and remained related to postural position. Since the IH syndrome has been mostly self-limited, conservative treatment with absolute bedrest, adequate hydration therapy, and NSAIDs were given. Two weeks after beginning treatment the orthostatic

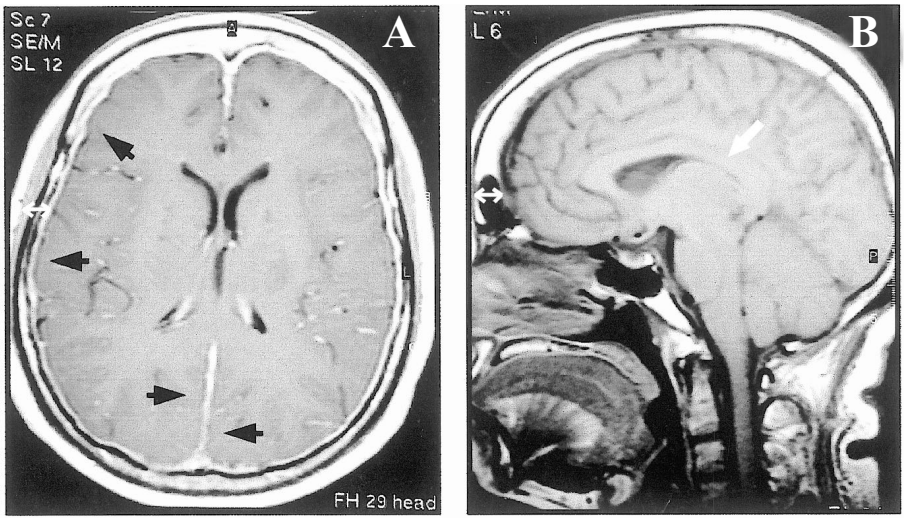

Fig. 1 (A) Diffuse meningeal enhancement with thickening of dura and cerebral falx (black arrows) on contrast-enhanced T1-weighted MR image. (B) T1-weighted sagittal imaging demonstrated mild cerebellar tonsillar descent, low position of the corpus callosum with compression of third ventricle (white arrow) and crowding of posterior fossa structures, flattening of the pons, and small prepontine, superior cerebellar, and inferior cerebellar cisterns.

headaches gradually subsided and no more bed rest or medications were needed. A follow-up brain MRI at 2 months after treatment revealed diminished meningeal enhancement (Fig. 2A). In addition, evidence of reduced amounts of subdural effusion, diminished displacement of the corpus callosum, as well as restoration of cerebellar tonsils to a position just above the foramen magnum were also found (Fig. 2B). Six months after the episode, meningeal enhancement had fully resolved as shown on follow-up MRI (Fig. 3).

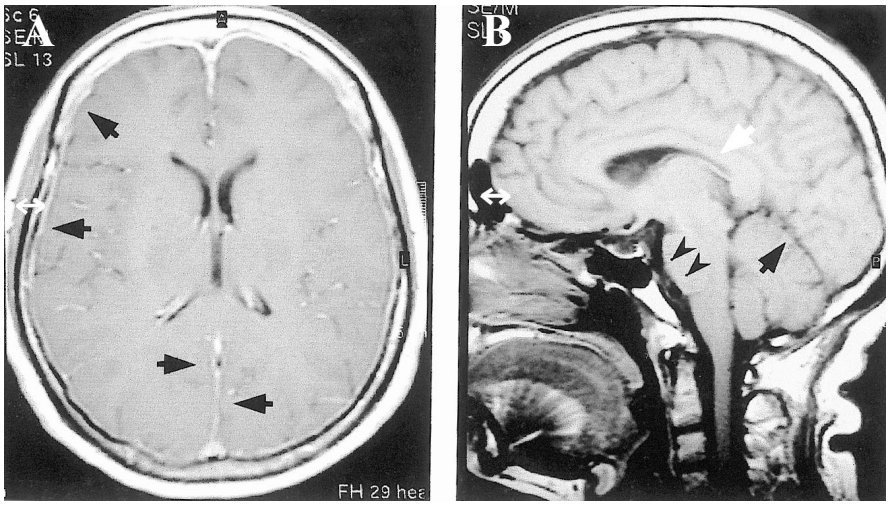

Fig. 2 Two-month follow-up magnetic resonance imaging. (A) Partial resolution of meningeal enhancement (black arrows). (B) Improvement of pontine flattening (arrow heads), restoration of the superior cerebellar (black arrow) and inferior cerebellar cisterns, as well as recovery of displaced corpus callosum (white arrow) were seen.

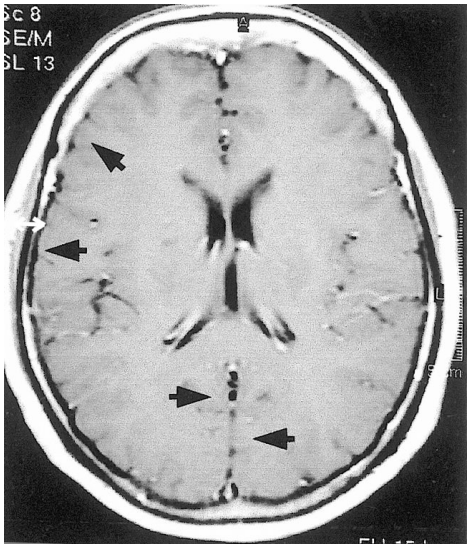

Fig. 3 Six-month follow-up magnetic resonance imaging revealed full resolution of abnormal meningeal enhancement (black arrows).

DISCUSSION

It is well known that headaches are one of the most common symptoms in patients with IH syndrome. The site of the headaches may be at the frontal, vertex, occipital regions, or the whole head. The characteristics are often varied, either throbbing or tension in nature, and are usually not relieved by analgesics alone.[5] Importantly, the head pain also varies in intensity, with associated clinical presentations such as dizziness, nausea, vomiting, and tinnitus.[8] Our patient presented with throbbing pain at the vertex in association with neck tightness, dizziness, nausea, and vomiting. In addition, her headaches were relieved quickly on recumbence. Therefore, a detailed search of causes underlying orthostatic headaches is warranted.

In IH syndrome, low CSF pressure in the absence of a spinal block is an important diagnostic

clue. The opening CSF pressure in our patient was 30 mm, a level below normal ranges at the lumbar level. Nevertheless, low CSF pressure is not always present in patients with IH syndrome since Mokri et al. located spinal pressure between 65 and 140 mm CSF in five out of 26 IH patients.[7] On the other hand, augmentation of headaches with jugular compression, which actually increases the intracranial pressure, suggests that headaches in IH patients are not solely related to the intracranial pressure.[9] Grant and his colleague proposed that headache due to CSF leakage were caused by reduced CSF volume rather than lowered CSF pressure.[10] As a result of the reduction of CSF volume, venous dilation or distortion of the supporting structures of the brain may appear.[11] Accordingly, the source of headaches in patients with IH may come from painful venous dilatation or brain descent with traction of pain-sensitive structures.

Spontaneous IH syndrome, originally described by Schaltenbrand,[12] was thought to result from overt CSF absorption or occult CSF leakage from small dural tears. Rupture of perineural or epineural cyst, tearing of spinal meningeal diverticulum or nerve sheath all account for cryptic leakage of the spinal fluid.[3-5,13] Of particular importance, radionuclide cisternography or CT-myelogram can be helpful in detecting the CSF leakage sites.[3,14] In our patient the cisternography demonstrated no evidence of CSF leakage, however, slow isotope ascent and reduced hemispheric radionuclide activities were observed. These findings suggest either a possible CSF hyperabsorption state or an existing CSF leakage too small to be detected using the current studies.

Characteristic MRI findings of spontaneous IH syndrome include subdural fluid collections, brain sagging,[7] and diffuse pachymeningeal contrast enhancement.[6] Formation of subdural hematomas or hygromas have been frequently observed in patients with sustained IH syndrome.[4] These vascular complications may enrich clinical manifestations and contribute to, at least in part, the occurrence of postural headaches. Presumably, these are caused by ruptures of bridging veins when the brain pulls away from the dura as a result of decreased CSF volume.[4] In our patient, diminished amount of subdural effusion was evident after proper management and this substantiated the role of CSF volume. The descent of the brain has been described as inferior displacement of the optic chiasma and iter, flattening of the pons, effacement of the prepontine, superior and inferior cerebellar cisterns, and caudal displacement of the pons and cerebellar tonsils.[15] In some patients, the descent of the brain can be measured on the midsagittal plane of an MRI.[16] First reported in 1991, the characteristic MRI findings for patients with IH syndrome included diffuse, even, uninterrupted enhancement involving the pachymeninges with thickened dura on contrast-enhanced T1-weighted and slight hyperintense on T2-weighted imaging. The mechanism of meningeal enhancement may be due to inflammation of the pachymeninges secondary to reversible disturbance of the choroid plexus,[16] dural venous dilatation secondary to reduced CSF volume,[15] or fibrocollagenous proliferation of the leptomeninges.[17] Resolution of imaging abnormalities has been emphasized after proper treatment and used to be in accordance to clinical improvement.[4,7,16,18] In our patient, a follow-up MRI performed 2 months after treatment showed persistent pachymeningeal enhancement when her headaches were totally abated. Complete resolution of pachymeningeal abnormalities was secured between the second and the sixth month in the symptom free stage. Since striking reduction in the degree of dural enhancement can be seen in days or weeks with regard to the theories of pachymeningeal inflammation and dural venous dilatation,[15,16] the longer period of meningeal enhancement reflects the chronic nature of the underlying pathology. Thus, such an extended interval of dural enhancement as seen in our patient may suggest proliferative changes of the leptomeninges, which is a more plausible mechanism responsible for this delayed reversible phenomenon.

REFERENCES

1. Bell WE, Joynt RJ, Sahs AL. Low spinal fluid pressure syndromes. Neurology 1960;10:512-21.
2. Lasater GM. Primary intracranial hypotension: the low spinal fluid pressure syndrome. Headache 1970;10:63-6.
3. Marcelis J, Silberstein SD. Spontaneous low cerebrospinal fluid pressure headache. Headache 1990;30: 192-6.
4. Rando TA, Fishman RA. Spontaneous intracranial hypotension: report of two cases and review of the literature. Neurology 1992;42:481-7.
5. Silberstein SD, Marcelis J. Headache associated with

abnormalities in intracranial structure or pressure. In: Dalessio DJ, Silberstein SD, eds. Wolff's Headache and Other Head Pain. 6th Ed. New York: Oxford University Press, 1993:438-61.
6. Mokri B, Krueger BR, Miller GM, Piepgras DG, Rochester MN. Meningeal gadolinium enhancement in low pressure headaches. Ann Neurol 1991;30:294-5.
7. Mokri B, Piepgras DG, Miller GM. Syndrome of orthostatic headaches and diffuse pachymeningeal gadolinium enhancement. Mayo Clin Proc 1997;72:400-13.
8. Lybecker H, Djernes M, Schmidt JF. Postdural puncture headache (PDPH): onset, duration, severity, and associated symptoms; an analysis of 75 consecutive patients with PDPH. Acta Anesthesiol Scand 1995;39:605-12.
9. Raskin NH. Lumbar puncture headache: a review. Headache 1990;30:197-200.
10. Grant R, Condon B, Hart I, Tesadale GM. Changes in intracranial CSF volume after lumbar puncture and their relationship to post-LP headache. J Neurol Neurosurg Psychiatry 1991;54:440-2.
11. Mokri B, Hunter SF, Atkinson JL, Piepgras DG. Orthostatic headaches caused by CSF leak but with normal CSF pressures. Neurology 1998;51:786-90.
12. Schaltenbrand G: Neure anshauungen zur pathophysiologie der liquorzirkulation. Zentrabl Neurorchir 1938;5:290-300.
13. Schievink WI, Torres VE. Spinal meningeal diverticula in autosomal dominant polycystic kidney disease. Lancet 1997;349:1223-4.
14. Benamor M, Tainturier C, Graveleau P, Pierot L. Radionuclide cisternography in spontaneous intracranial hypotension. Clin Nucl Med 1998;23:150-1.
15. Fishman RA, Dillon WP. Dural enhancement and cerebral displacement secondary to intracranial hypotension. Neurology 1993;43:609-11.
16. Pannullo SC, Reich JB, Krol G, Deck MD, Posner JB. MRI changes in intracranial hypotension. Neurology 1993;43:919-26.
17. Good DC, Ghobrial M. Pathologic changes associated with intracranial hypotension and meningeal enhancement on MRI. Neurology 1993;43:2698-700.
18. Hochman MS, Naidich TP, Kobetz SA, Fernandez-Maitin A. Spontaneous intracranial hypotension with pachymeningeal enhancement on MRI. Neurology 1992;42: 1628-30.

原發性顱內低壓症：可逆性硬腦膜顯影增強與腦降之磁振造影表現

曾昱龍 張永義 藍旻瑜 吳秀善 劉家壽

一位56歲的女性，因嚴重的姿勢性頭痛合併噁心嘔吐前來求診，腰椎穿刺檢查發現腦脊髓液壓明顯下降，臨床上診斷爲低顱內壓症候群。第一次的腦部磁振造影發現廣泛性的腦膜顯影以及腦部下墜，然放射性同位素腦池攝影未能發現明顯的脊髓液滲漏。患者的頭痛經兩週治療後已完全恢復，但兩個月後追蹤的腦部磁振造影發現腦膜顯影僅有部分改善，直到6個月後才完全消失。由此可見罹患自發性低顱內壓症候群之患者，其腦膜顯影的現象是可逆性的，我們推論此種延遲性腦膜顯影消失的變化可能與腦膜的增生有關。(長庚醫誌 2003;26:293-8)

關鍵字：低顱內壓症候群，姿勢性頭痛，腦脊髓液，磁振造影。

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